256* Patients with cystic fibrosis can preserve peak working capacity, even if lung function is substantially impaired

L. Lannefors[1], U. Dennersten[1], M. Ortfelt[2], H. Johansson[3], K. Hellberg[1], P. Höglund[4], C.-G. Löfdahl[1]. [1]*Lund University Hospital, Pulm Med, Lund, Sweden;* [2]*Stockholm University, CF-centre, Stockholm, Sweden;* [3]*Uppsala Univ Hospital, CF-centre, Childrens Hospital, Sweden;* [4]*Lund University Hospital, Competence Centre for Clinical Research, Lund, Sweden*

Background: Swedish Cystic Fibrosis (CF) care follows international guidelines in general. However, individually tailored moderate intense physical handling in infants and thereafter physical exercise has been the primary base for the airway clearance therapy for >25 years. The aims of this study were to describe the Peak Working Capacity (PWC) in a Swedish multi CF centre population, especially in the subgroup with substantially impaired lung function, and to evaluate PWC rate of change over a three year period.
Methods: Simultaneous measurements of PWC and FEV_1 were collected from medical records and presented as % of predicted (%p) in all patients at three national CF centres, comprising 74% of all Swedish CF patients. All patients independent of age had been equipped with individually tailored and frequently optimized treatment programs including physical exercise since 1984 or the day of diagnosis.
Results: Of all 205 eligible CF patients $\geqslant$7 years old 191 (93%) were included. Mean age was 20 (range 7–60) years. Mean (SD) FEV_1 was 77(25) %p and PWC 100(26) %p (females 105 and males 94%p). Individuals with $FEV_1 \leqslant 50$ %p showed mean PWC 75(23) %p. Mean annual rate of change of PWC %p was −1.0(6.6), $p = 0.071$. Annual rate of change was dependent on annual FEV_1 %p rate of change, $p < 0.0001$, but independent of age, and of FEV_1 %p at start, $p = 0.395$.
Conclusion: PWC can be well maintained over time in CF patients, even if lung function is substantially impaired. A CF care package with a physiotherapy regimen including physical exercise from the day of diagnosis could possibly explain these results.

257 The provision of exercise assessment across UK CF centres

A.R. Morris[1], A.M. Collins[2], E. Spencer[1], E. Lloyd[1], A. Ashish[1], M.J. Ledson[1], M.J. Walshaw[1]. [1]*Liverpool Heart and Chest Hospital NHS Foundation Trust, Regional CF Centre, Liverpool, United Kingdom;* [2]*Royal Liverpool and Broadgreen University Hospitals NHS Trust, Liverpool, United Kingdom*

Regular assessment of exercise capacity in CF is a useful tool to aid exercise prescription and monitor changes in disease state. In particular, aerobic tests measuring peak exercise capacity and sub-maximal exercise responses are especially useful in highlighting changes in aerobic fitness, functional ability and any subsequent impact on quality of life. We aimed to assess which exercise tests, if any, are used in UK CF centres.
We surveyed 40 UK centres (19 adult, 21 paediatric) units using a structured questionnaire asking about the timing and use of exercise testing for their CF patients. Only 35 centres (88%) routinely measured exercise capacity in their patients. Peak exercise capacity (VO2 Peak) was measured in 32 (91%) and sub-maximal or steady state exercise capacity (VO2 MSS) in 21 (60%), with 13 centres (37%) using both tests. Of those centres measuring peak exercise, patients were assessed via a shuttle test in 20 (63%), the Chester step test in 7 (22%) and via cycle ergometer in 5 (16%). Sub-maximal values were assessed via a 6-min walk in 13 (62%) centres, YMCA 3-min step test in 5 (24%), treadmill in 2 (10%) and cycle ergometer in 1 (5%). As regards timing, 28 centres (80%) assessed exercise capacity routinely at annual review (14 of these at no other time), 15 (43%) routinely during an inpatient stay, 10 (29%) due to clinical need and 7 (20%) prior to exercise prescription.
Standardised tests to assess exercise capacity in CF patients would improve inter-centre communication, especially during transition. We are now designing a study to ascertain which of these tests best measures exercise capacity in our patients.

258 Peak work capacity and utilization of healthcare resources in adult CF patients

A. Ashish[1], A. Morris[1], J. McShane[1], M.J. Ledson[1], M.J. Walshaw[1]. [1]*Liverpool Heart and Chest Hospital, Liverpool, United Kingdom*

Peak work capacity (PWC%) (corrected for lung function) measures physiological reserve and can be used to estimate the patient's fitness, which in turn may reflect the burden they place on healthcare resource use. Currently, the most commonly used prognostic indicator in CF patients is the FEV1, which is inversely related to treatment burden and healthcare utilisation. We have compared the relationship between PWC%, FEV1, and healthcare resource utilisation in a group of 75 adult CF patients (mean age 27 years [range 20–48], 45 males) attending our large adult clinic.
PWC% (measured during a phase of clinical stability) was correlated with the best FEV1 (% predicted over the preceding year), inpatient days, and length of stay and outpatient visits over a 4 year period. Kendall's rank correlation was used to analyse the association between these variables.
Although PWC% correlated concordantly with FEV1 ($p < 0.001$), it had a stronger correlation than FEV1 with the number of admissions (0.0008 v 0.004), inpatients days (0.0005 v 0.007), and outpatient attendances (0.02 v 0.10) respectively.
Thus study shows that Peak Work Capacity is a sensitive indicator of healthcare resource utilization in CF patients, and its measurement should be used more routinely in CF centers. It may prove to be a useful adjunct to the currently available prognostic indicators in this increasingly common chronic condition.

259 Reliability of cycle ergometer tests and the LifeShirt in children with CF

L. Kent[1], B. O'Neill[1], J. Murray[2], A. Reid[2], G. Davison[1], A. Nevill[3], J.S. Elborn[4,5], J. Bradley[1,5]. [1]*University of Ulster, Jordanstown, United Kingdom;* [2]*Royal Belfast Hospital for Sick Children, Belfast, United Kingdom;* [3]*University of Wolverhampton, Wolverhampton, United Kingdom;* [4]*Queen's University Belfast, Belfast, United Kingdom;* [5]*Belfast City Hospital, Belfast, United Kingdom*

Introduction: There is limited research on the use of exercise tests outside the clinical laboratory setting in children with CF.
Aims: To assess reliability and acceptability of (a) cycle ergometry tests, and (b) using the LifeShirt to record cardiorespiratory responses during cycle ergometry tests, in children with CF.
Methods: Children with CF and stable lung disease wore the LifeShirt during 2 cycle ergometry tests (intermittent sprint test (IST) measuring muscle power; continuous incremental test (CIT) measuring cardiorespiratory fitness) on 2 occasions 7–8 days apart. Acceptability data were collected by questionnaire.
Results: 10 children with CF completed the study: (5M:5F), 9(2) y, FEV_1: 83(14)%. Peak power (IST) and peak work capacity (CIT) were reliable (bias $p > 0.05$, CV < 10%). Tidal volume, ventilation, respiratory rate, fractional inspiratory time, heart rate and SpO_2 recorded by the LifeShirt demonstrated no significant bias ($p > 0.05$). Respiratory rate, fractional inspiratory time, heart rate and SpO_2 demonstrated acceptable CVs (<10%). Acceptability questions indicated that cycle ergometer tests and the LifeShirt were mainly acceptable to the children.
Conclusion: This is the first study to report reliability of cycle ergometry in children with CF. The LifeShirt may be useful for simultaneous measurement of respiratory rate, fractional inspiratory time, heart rate and SpO_2 outside the normal clinical laboratory setting, however, there is concern over volume components of breathing in children with CF.